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Case Report

Pancreatic pseudopapillary tumor in association with colonic polyposis

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Abstract

We present a case of a female patient with familial adenomatous polyposis (FAP) and a history of pseudopapillary pancreatic tumor two years earlier. FAP is accepted to be a rare precancerous syndrome characterized by hundreds to thousands of colorectal adenomatous polyps with a variety of extraintestinal manifestations. Solid pseudopapillary tumor (SPT) is an uncommon exocrine pancreatic neoplasm. This rare pancreatic tumor should be suspected in FAP patients especially if the patient has abdominal complaints. The development of two rare entities in the same patient may suggest an association and research for a certain genetic basis might be indicated.

Keywords: Familial adenomatous polyposis, pancreatic tumor, extraintestinal, colonoscopy, malignancy.

INTRODUCTION

It has long been recognized that familial adenomatous polyposis (FAP) is an autosomal dominant disease with a prevalence of about 1 in 5000-10000 people caused by germline mutation of the adenomatous polyposis coli (APC) gene located on chromosome 5q21. As a de novo syndrome FAP may be seen without positive family history. APC mutations result in an increased level of intracellular b-catenine protein leading to upregulation of T cell factor responsive genes needed for proliferative and neoplastic changes of epithelial cells. FAP is characterized by the development of uncountable number of colorectal adenomatous polyps. Patients with FAP have a 100% chance to develop colorectal malignancy, usually before the fourth decade, if colectomy is not performed (Half et al., 2009; Laurent et al., 2011; Maire et al., 2002).

A variety of other extracolonic lesions have been reported linked to FAP, including benign osteomas, desmoid tumors, congenital hypertrophy of retinal pigment epithelium, fundic gland polyposis, nasopharyngeal angiofibromas and central nervous system lesions. In addition, malignancies of duodenum, pancreas, biliary tract, stomach, liver, thyroid and endocrine neoplasia are well described associated with FAP (Lee et al., 2004; Campos et al., 2003; Righetti et al., 2011). Solid pseudopapillary tumors (SPTs) of pancreas (Frantz tumor) are a rare, usually benign tumor of the pancreas with a low malignant potential. This is in contrast to the usually malignant pancreatic tumors seen in association with adenomatous polyposis. These tumors are mostly seen in young female in the second or third decade. Rarely, these tumors may be seen in children. The most common clinical presentation is a palpable abdominal mass and an uncharacteristic abdominal pain (Kamat et al., 2008; Munding et al., 2001).

We report here a case of 14-year-old female with a history of pseudopapillary tumor of the pancreas that was found to have adenomatous polyposis syndrome.

The importance of accurate diagnosis and treatment is emphasized.

Case Report

A 14 year-old girl presented with progressive fatigue, intermittent loose stools, blood mixed in her stool and vague abdominal pain for 3 months. She had a history of laparotomy 2 years earlier due to progressive abdominal pain and a documented diagnosis of pseudopapillary

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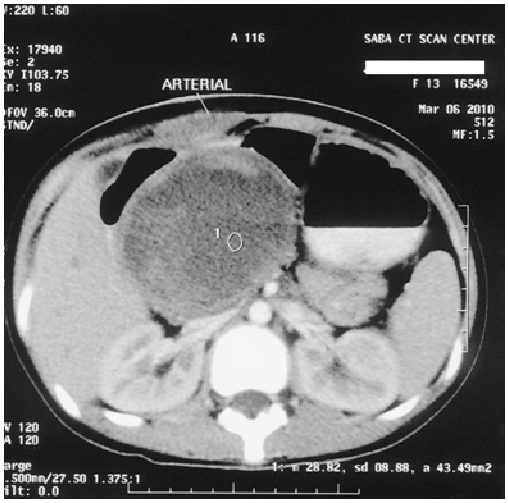

Figure1. Abdominal CT showing pancreatic mass

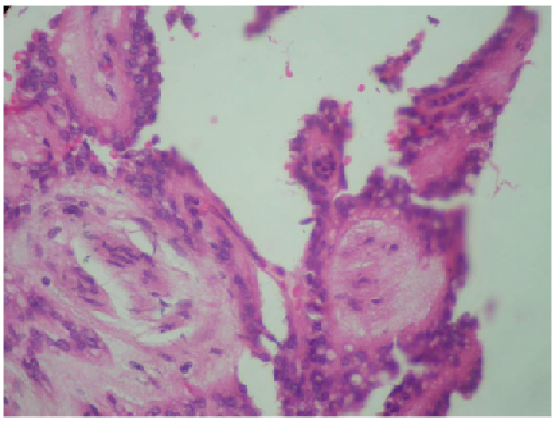

Figure 2. Histopathological view of the pancreatic mass showing myxoid connective tissue and small cells with bland-looking nuclei making a pseudopapillary pattern of growth, H and E staining 400X

pancreatic tumor. For additional information, we sought the patient's previous medical records. Her abdominal computed tomography (CT) had shown a large encapsulated pancreatic mass with solid and cystic components (Figure1). Histopathological investigation had revealed necrotic changes and the presence of hypercellularity with polygonal nuclei, inconspicuous nuclei, a pseudopapillary and cystic pattern with proliferation of myxoid connective tissue arranged in solid sheets around tiny blood vessels without mitotic figures and any vascular invasion (Figure 2). The tumor findings were: CD10 (+), synaptophysin (+), chromogranin A(+),vimentin(+), neuron-specific enolase (NSE)(+) and calretinin(-).

The patient was well during about 18 months after tumor resection and there was no need to chemotherapy. Her follow-up abdominal CT 1 year after surgery revealed no mass lesion and any other abnormality. Family history was unremarkable. There was nothing in her history or physical examination to suggest sexual abuse. On physical examination, she was skinny, pale and emaciated with a laparotomy scar. Hemoccult test was positive, and the examinations of the stool for ova and parasite were negative. She had negative stool and blood cultures. Blood analysis showed anemia. Liver and kidney function tests were normal. The patient had normal vital signs except for a heart rate of 120/min. Lab tests revealed decreased Hb 7g/dL (normal 11-14), total protein 4g/dL (normal 5.5-8.5) and serum albumin 2g/dL (normal 3.5-5.5). Prothrombin time, partial thromboplastin time, and erythrocyte sedimentation rate were normal. Abdominal CT and ultrasonography showed rectal and secal wall edema. According to clinical and laboratory picture, our first suspicion was that the patient might be

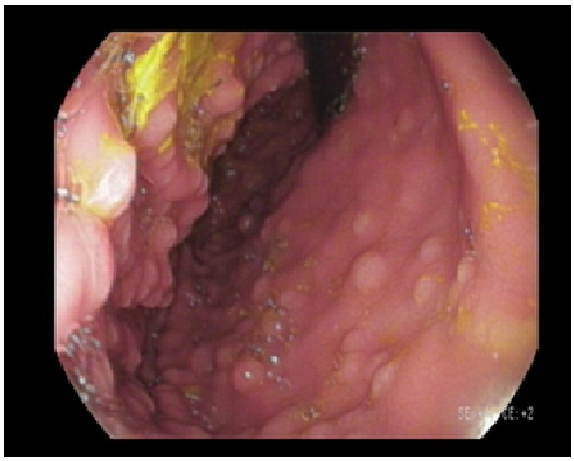

Figure 3. Endoscopic view of gastric fundus revealing fundic gland polyps(5-10 mm)

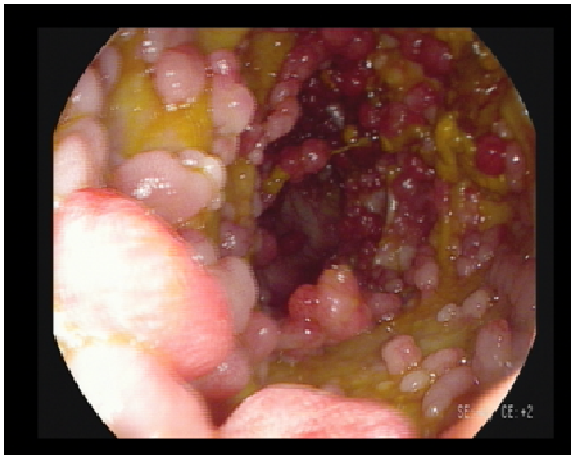

Figure 4. Colonoscopic view into the descending colon revealing multiple polyps (5–20 mm)

afflicted with Inflammatory Bowel Disease (especially Crohn's disease).

Esophagogastroduodenoscopy revealed multiple nodular lesions in the gastric antrum, body and fundus (measuring 0.5-1cm) (Figure3), gastric antral mucusa was diffusely nodular and erythematous. Interestingly, the colonoscopy upto cecum revealed multiple polyps throughout the colon (measuring 0.5-2 cm), in the rectum, sigmoid colon, descending, transverse and ascending colon (Figure 4). Four pedunculated rectal polyps were removed by snare polypectomy and sent to pathology. Histopathology reporting of the colonic specimens was adenomatous polyp with low grade dysplasia (Figure 5). Brain imaging and ophtalmologic examination revealed no abnormality. Small bowel follow through with oral barium and capsule endoscopy detected no polyps in the small intestine. Genetic evaluation revealed a mutation in the APC gene on chromosome 5. Total colectomy with ileoanal anastomosis, postsurgical surveillance, and family screening were recommended.

DISCUSSION

We report an association of FAP with pancreatic pseudopapillary tumor in an adolescent girl. Given the rarity of this pancreatic tumor, its association with colonic polyposis seems noteworthy.

Extracolonic benign and malignant neoplasms have been described in patients with adenomatous polyposis syndromes. Reported pancreatic lesions associated with FAP are quite rare. One case of pancreatic glucagonoma has been described. Other pancreatic lesions reported in association with FAP include intraductal papillary

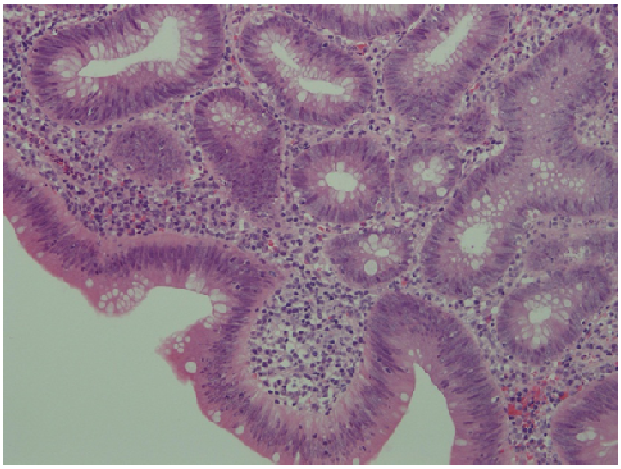

Figure 5. Mildly dysplastic epithelium lined the surface of colonic polype, H and E staining 400X

mucinous pancreatic neoplasms, pancreatic duct adenomas, pancreatic intraepithelial neoplasia and islet cell tumors. Due to the rarity of these and other types of malignancies reported in association with FAP, true linkage will be difficult to prove(Lee et al., 2004; Campos et al., 2003). Abraham et al(2001) described molecular alterations in the APC/beta-catenin pathway in a patient with pancreatoblastomas who had the FAP mutation. There are a few reports describing carcinoid tumors in association with FAP. No known genetic basis exists explaining the link between FAP and carcinoid tumor (Camp et al., 2004).

In our literature review, we found only two previous report of pancreatic pseudopapillary tumor in association with colonic polyposis. In a a study by Ruo et al(2002) evaluating the clinical outcome of FAP patients, this tumor was diagnosed in histopathological evaluation in a 43-year old female patient after pancreaticoduodenal surgery for periampullary neoplasm. In another report, a young 29-year old man with familial adenomatous polyposis was described in association with Frantz tumor of the pancreas (Le Borgne et al., 1997).The interesting findings in our case is that adenomatous polyposis was diagnosed after pancreatic tumor and there was no family history of colonic polyposis.

Solid pseudopapillary tumor of the pancreas accounts for 1-2% of exocrine pancreas tumors. Frantz first described this tumor in 1959 as a “papillary tumor of the pancreas”. The primary definition did not determine its benign or malignant entity. This uncommon, typically benign tumor seems to have a predilection for young girls and women between the 2nd and 3rd decades of life. They are also called Gruber-Frantz tumor, Frantz tumor, solid- cystic and solid- papillary neoplasm of the pancreas. The tumor usually presents with minimal symptoms such as nonlocalized abdominal pain or discomfort. The abdomen is usually nontender on palpation; there are usually no abnormalities in laboratory tests. SPTs should be added to the list of extracolonic tumors that can occur in patients with FAP. These tumors should be considered in cases of nonspecific persistent abdominal complaints, acute pancreatitis, diabetes, or steatorrhea in patients with FAP (Kamat et al., 2008; Munding et al., 2001; Le Borgne et al., 1997). There is not enough information on the genetic basis of these lesions. The rarity of these tumors makes it appear that the association is more than a simple coincidence.

Screening for colorectal and duodenal cancer (gastrointestinal endoscopy) is necessary in patients with FAP; because of their scarcity, routine screening for pancreatic tumors seems unwarranted. However, we recommend that in known cases of FAP with certain abdominal complaints, special consideration should be given to the rare pancreatic tumors.

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